Anaesthetic management of a parturient with moyamoya disease

Dutta B, Dehran M, Sinha R

ABSTRACT

Moyamoya disease is an uncommon progressive cerebral vasculopathy that is more frequently seen in the Asian population. It has been described sporadically in other parts of the world. Proper knowledge of the pathophysiology and institution of appropriate perioperative measures improves patient prognosis. We report a case of moyamoya disease for emergency Caesarean section at 37 weeks of gestation. Epidural anaesthesia was administered using 0.5 percent bupivacaine and 50 µg fentanyl. A stable haemodynamic state was maintained using left lateral uterine displacement, intravenous crystalloids and ephedrine boluses. The patient suffered no neurological deficit and was discharged with a healthy baby four days after the surgery.

Keywords: Caesarean section, epidural anaesthesia, moyamoya disease

Singapore Med J 2011;52(6):e108-e110

INTRODUCTION

Moyamoya disease (MMD) is a rare, progressive cerebrovascular occlusive disease of the Circle of Willis and its feeding arteries. Pregnant patients with MMD are at a higher risk of neurological deterioration due to cerebral ischaemia or haemorrhage.[1] Delivery by Caesarean section is recommended, as normal delivery can be deleterious. Both general and regional anaesthetic techniques have been practiced for these patients. We report successful anaesthetic management of lower segment Caesarean section in a term parturient with MMD.

CASE REPORT

A 27-year-old, 52-kg primigravida with MMD was admitted at 37 weeks of gestation for elective Caesarean section. However, emergency lower segment Caesarean section was planned due to early onset of labour pains. The patient was a known case of MMD and had spontaneous left gangliocapsular bleed in the fifth month of gestation. She showed good recovery without any neurological deficit with conservative management. During preoperative examination, she was alert and oriented, with mild cognitive dysfunction and intermittent short-term memory loss. She had good exercise tolerance with a heart rate of 78 beats/min and a non-invasive blood pressure (NIBP) of 122/86 mmHg. Her mean arterial pressure (MAP) was 98 mmHg. Neurological examination revealed no signs of cerebral ischaemia. She was not on any medication. Her haematological investigations were within normal limits. Magnetic resonance (MR) imaging of the brain done four months earlier showed left gangliocapsular bleed with intraventricular extension, mild ventricular dilatation with periventricular ooze, a lack of normal flow void in the right internal carotid artery, a narrowed right middle cerebral artery and multiple suprachiasmatic flow voids.

Epidural anaesthesia was planned. The patient was premedicated with ranitidine 50 mg intravenously and 0.3 M sodium citrate 30 ml orally. In the operating room, standard monitoring, including electrocardiography, NIBP and pulse oximeter were used. A 16 G intravenous cannula was inserted and oxygen was administered by face mask. After preloading with lactated Ringer's solution (15 ml/kg), an epidural catheter was placed in the second and third lumbar interspace in the left lateral position, with 4 cm of the catheter in the epidural space. The patient was then turned supine with a slight left lateral tilt. Intravascular and intrathecal placement of the catheter was ruled out with an epidural test dose of 3 ml 2% lignocaine-adrenaline (1:200,000). Sensory analgesia till the sixth thoracic dermatome was achieved with incremental doses of 0.5% bupivacaine (18 ml) combined with 50 µg fentanyl before surgery commenced.

Oxytocin infusion was started after delivery of the placenta, following which there were two episodes of hypotension (MAP < 20% from the baseline) to 72 mmHg at ten minutes and 76 mmHg in 15 minutes. These episodes were corrected by increasing the rate of crystalloid infusion and two boluses of intravenous ephedrine (6 mg each). The duration of surgery was one hour, and blood loss was approximately 400 ml. Normothermia was achieved with stable operating room temperature, warm intravenous fluids and a warming blanket. Verbal contact was maintained throughout, and bilateral handgrip was checked intermittently. The patient did not show any signs

Department of Anaesthetics, Peterborough and Stamford Hospitals, NHS Foundation Trust, Bretton Gate, Peterborough PE3 9GZ, UK

Dutta B, MD
Speciality Registrar

Department of Anaesthesiology and Critical Care, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India

Dehran M, MD
Professor

Sinha R, MD
Associate Professor

Correspondence to:
Dr Bhaskar Dutta
Tel: (44) 772 996 3759
Fax: (44) 1773 678 533
Email: dr_dbhaskar@yahoo.com

or symptoms of cerebral ischaemia. Postoperatively, the patient was monitored in a high-dependency area for 24 hours, where oxygen therapy was continued. Analgesia was maintained with 2-mg epidural morphine after every 12 hours. The postoperative period was uneventful, and the patient was discharged four days after surgery with a healthy baby boy.

DISCUSSION

MMD is characterised by progressive occlusion of the cerebral vasculature, particularly the Circle of Willis and its feeding arteries. Moyamoya means 'puff of smoke' in Japanese, which describes the angiographic appearance of the abnormal vascular collateral networks adjacent to the stenotic vessels.[1] MMD is hereditary (autosomal dominant)[2] with a specific gene locus, q25.3, on chromosome 17.[3] The prevalence and incidence is approximately 3.16 cases and 0.35 cases per 100,000 people, respectively with a female-to-male ratio of 1.8:1.[4] Mortality rates are approximately 10% in adults and 4.3% in children. MMD is associated with other diseases, including thyrotoxicosis, sickle cell anaemia, Down's syndrome, coarctation of the aorta and hypertension, which warrant consideration due to their implications during anaesthesia and surgery. Pharmacotherapy in MMD patients includes the use of antihypertensives, anticoagulants or antiplatelet agents (e.g. aspirin). Various surgical procedures have also been used, e.g. superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis, encephaloduroarteriosynangiosis and encephaloduroarteriomyosynangiosis. The prognosis is best after the STA-MCA surgery, with an immediate increase in blood supply.

Multiple strokes or transient ischaemic attacks (TIA) may lead to mental deterioration and can be fatal due to intracerebral haemorrhage (ICH), especially at term or during labour. ICH can be confused with pregnancy-induced hypertension, as transient hypertension and proteinuria can complicate ICH. A non-contrast computed tomography or MR imaging is helpful to differentiate between the two conditions. The basic aim of anaesthesia is to maintain the balance between cerebral metabolic oxygen consumption rate ($CMRO_2$) and cerebral blood flow (CBF) in order to prevent any neurologic morbidity. The anaesthetic technique should ensure normotension, normocapnia and normothermia in the perioperative period.

Carbon dioxide is a potent modulator of cerebrovascular tone. Hypocarbia is deleterious due to marked reductions in regional CBF at $PaCO_2 < 29$ mmHg.[5] Hyperventilation leads to vasoconstriction of moyamoya vessels,[6] which causes a significant decrease in regional cortical blood flow, diffuse slowing of electroencephalogram (EEG) and clinical cerebral ischaemia.[7] After hyperventilation subsides, there is a 'steal' from the moyamoya vessels to the dilated cortical vessels, resulting in long-lasting ischaemia.[8] Hypercapnia can also be dangerous, as Tatemichi et al demonstrated in their study of reduced hypercapnic vasoreactivity in adults with MMD.[9] Normothermia should be maintained to optimise CBF, as hypothermia precipitates vasospasm.[10] CBF is lesser in MMD patients than in healthy individuals.[11] It has been observed that the mean velocity of CBF starts decreasing in proportion to reductions in MAP, leading to cerebral hypoperfusion. Therefore, maintenance of MAP is important for optimal CBF.[6]

Elective Caesarean section is recommended to avoid deleterious effects of pain, hyperventilation and bearing down during labour.[1] However, successful vaginal deliveries under spinal and epidural analgesia have also been reported.[12] Anaesthetic approach in these parturients undergoing lower segment Caesarean section has not been standardised in the literature. General anaesthesia has many disadvantages apart from risks of gastric aspiration and neonatal depression. Laryngoscopy, tracheal intubation, extubation and inadequate depth of anaesthesia can all lead to an increase in $CMRO_2$,[8] and may result in ICH due to rupture of the fragile network of collaterals or defined saccular aneurysms under increased haemodynamic stress.[6] Halogenated anaesthetics produce dose-dependent decrease in peripheral vascular resistance and cerebrovascular tone,[13,14] and can abolish cerebrovascular autoregulation.[8] The potential advantage of general anaesthesia is decreased in $CMRO_2$, which may confer some protection against ischaemia. It also allows for optimal control of ventilation. Propofol anaesthesia has been successfully used for Caesarean section in a patient with MMD.[15] Continuous propofol infusion reduces the hypertensive response to laryngoscopy and intubation.[15,16]

Neuraxial blockade allows for continuous monitoring of cerebral function.[12] Epidural analgesia prevents hypertension, hyperventilation and hypocarbia associated with labour.[17] However, neuraxial blockade may lead to hypotension due to sympathetic block that may lower cerebral perfusion pressure, leading to cerebral ischaemia.[1] Epidural anaesthesia is associated with gradual onset of sympathetic block with decreased incidence of hypotension, and is therefore favourable for patients with MMD. Subarachnoid block causes a sudden

fall in MAP, which is undesirable in these patients, as it would complicate the already compromised cerebral circulation. There is a case report of convulsions and hemiparesis following subarachnoid block in a child with MMD;[18] hence, it was avoided in our patient. Our aim was to maintain MAP above 80 mmHg so that there was < 20% reduction from the preoperative MAP value. Preloading with left lateral tilt prevented a precipitous fall in blood pressure in our patient. Ngan Kee and Gomersall used continuous infusion of ephedrine at a dose of 100–500 μg/min for maintaining MAP.[1] We avoided ephedrine infusion in order to prevent an inadvertent rise in MAP, which could have been detrimental in this patient.

Intraoperative monitoring of CBF or cerebral ischaemia is a useful adjunct in MMD patients. CBF can be monitored either by ^{133}Xe technique or by measuring CBF velocity using transcranial Doppler, but there are limitations. ^{133}Xe poses radiation hazard whereas transcranial Doppler is effective in determining CBF velocity only in large vessels (e.g. middle cerebral and internal carotid arteries), which are already occluded or severely stenotic in MMD and thus, not very useful. Cerebral ischaemia can also be detected by EEG and near-infrared spectroscopy, which uses spectral analysis of reflected infrared light to measure cerebral oxygenation.[19] Although it is an excellent non-invasive monitor, prolonged preparation time is disadvantageous in emergency situations.[1] Since these specialised neurological monitoring techniques were not available at our emergency operating room, we aimed to keep the MAP around the preoperative levels, and maintained continuous verbal communication with the patient.

Management of postoperative analgesia deserves special mention in these patients, as postoperative pain may lead to the vicious cycle of anxiety-hyperventilation-hypocapnia, with resultant decrease in CBF. We used epidural morphine for the same, which provided adequate pain relief. Epidural local anaesthetics were avoided so that any inadvertent hypotension could be averted in the postoperative period. In conclusion, anaesthetic management in term parturients with MMD who undergo emergency Caesarean section should focus on the maintenance of adequate CBF, and the key to achieving this goal is maintenance of normothermia, normocapnia and normotension.

REFERENCES

1. Ngan Kee WD, Gomersall CD. Extradural anaesthesia for caesarean section in a patient with moyamoya disease. Br J Anaesth 1996; 77:550-2.
2. Mineharu Y, Takenaka K, Yamakawa H, et al. Inheritance pattern of familial moyamoya disease: autosomal dominant mode and genomic imprinting. J Neurol Neurosurg Psychiatry 2006; 77:1025-9.
3. Mineharu Y, Liu W, Inoue K, et al. Autosomal dominant moyamoya disease maps to chromosome 17q25.3. Neurology 2008; 70:2357-63.
4. Wakai K, Tamakoshi A, Ikezaki K, et al. Epidemiological features of moyamoya disease in Japan: findings from a nationwide survey. Clin Neurol Neurosurg 1997; 99 suppl 2:S1-5.
5. Tagawa T, Naritomi H, Mimaki T, Yabuuchi H, Sawada T. Regional cerebral blood flow, clinical manifestations, and age in children with moyamoya disease. Stroke 1987; 18:906-10.
6. Williams DL, Martin IL, Gully RM. Intracerebral hemorrhage and Moyamoya disease in pregnancy. Can J Anaesth 2000; 47:996-1000.
7. Kurokawa T, Chen YJ, Tomita S, Kishikawa T, Kitamura K. Cerebrovascular occlusive disease with and without the moyamoya vascular network in children. Neuropediatrics 1985; 16:29-32.
8. Soriano SG, Sethna NF, Scott RM. Anaesthetic management of children with moyamoya syndrome. Anesth Analg 1993; 77:1066-70.
9. Tatemichi TK, Prohovnik I, Mohr JP, et al. Reduced hypercapnic vasoreactivity in moyamoya disease. Neurology 1988; 38:1575-81.
10. Khan-Ghori SN, Murshid WR, Samarkandi AH, al-Salman M, Salih MA. Use of propofol and sevoflurane in moyamoya disease--case reports and literature review. Middle East J Anesthesiol 1999; 15:73-83.
11. Ogawa A, Yoshimoto T, Suzuki J, Sakurai Y. Cerebral blood flow in moyamoya disease. Part 1: Correlation with age and regional distribution. Acta Neurochir (Wien) 1990; 105:30-4.
12. Komiyama M, Yasui T, Kitano S, et al. Moyamoya disease and pregnancy: case report and review of the literature. Neurosurgery 1998; 43:360-9.
13. Eger EI 2nd, Smith NT, Stoelting RK, et al. Cardiovascular effects of halothane in man. Anesthesiology 1970; 32:396-409.
14. Stevens WC, Cromwell TH, Halsey MJ, et al. The cardiovascular effects of a new inhalation anesthetic, Forane, in human volunteers at constant arterial carbon dioxide tension. Anesthesiology 1971; 35:8-16.
15. Furuya A, Matsukawa T, Ozaki M, Kumazawa T. Propofol anaesthesia for cesarean section successfully managed in a patient with moyamoya disease. J Clin Anesth 1998; 10:242-5.
16. Gin T. Propofol during pregnancy. Acta Anaesthesiol Sin 1994; 32:127-32.
17. Fisher A, Prys-Roberts C. Maternal pulmonary gas exchange. A study during normal labour and extradural blockade. Anaesthesia 1968; 23:350-6.
18. Yasukawa M, Yasukawa K, Akagawa S, Nakagawa Y, Miyasaka K. Convulsions and temporary hemiparesis following spinal anaesthesia in a child with moyamoya disease. Anesthesiology 1988; 69:1023-4.
19. Harris DN. Near infra-red spectroscopy. Anaesthesia 1995; 50:1015-6.